WRIGHT (J.)

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A CASE OF PRIMARY LUPUS OF THE PHARYNX.

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THE recent flurry among the miracle-seeking members of our profession, caused by Koch's premature announcement, has been at least productive of the publication of records of cases of lupus.

For a number of years it was thought that lupus of the mucous membrane of the nose and mouth was always an extension, by continuity or contiguity, of a similar growth on the skin. In the last seven or eight years, however, reports of carefully studied cases have made it probable that a very large proportion (from 20 to 45 per cent.) of all cases of facial lupus really begin in the contiguous mucous membranes.[1] The painless character of the growth is an explanation of this former oversight on the part of clinical observers. Fifteen years ago, Von Ziemssen[2] rightly prophesied, and the prophecy was echoed by Lefferts,[3] that a more careful search would reveal many cases of lupus of the throat. Lennox Browne[4]

[1] Vide Bender's convincing articles, Vierteljahresschrift für Derm. und Syph., 1888, 15, p. 892, et alidem.

[2] Amer. transl. Encycl., 1876, vol. v, p. 852.

[3] Amer. Journ. Med. Sciences, April, 1878, vol. lxxv.

[4] Diseases of the Throat, 3d ed., p. 426.

has lately said that in a skin hospital he saw in a few weeks more cases of lupus of the nose and throat than he had previously seen in a throat-practice of twenty years. For a number of years nearly all the standard works and monographs have referred to the case mentioned by Von Ziemssen in 1876 as the only one reported in which the disease was primary and confined to the larynx. This statement is certainly erroneous, since Haslund,[1] in 1883, reported a case of lupus confined to the larynx, while Obertüschen[2] recorded a case in which the lupus first appeared in the larynx of an otherwise perfectly healthy individual; a year and a half later skin-lupus appeared on the nose, the laryngeal lesion having healed. Others more or less doubtful have been reported. Primary lupus of the tongue, though almost as rare as that of the larynx, has been observed by Fairlie Clarke[3] and Leloir.[4] Primary lupus of the soft palate and pharynx is more common, while the nasal mucous membrane is very frequently the starting-place of facial lupus. Of course, many of these cases are open to justifiable doubt in regard to the diagnosis; but in the light of the more recent publications of clinical experience one should hardly be satisfied with the declaration of Lefferts (*loc. cit.*), Störk,[5] and others, that they would decline to make a diagnosis of lupus of the throat without coëxisting skin-lupus. Such isolated

[1] Vierteljahressch. für Derm. und Syph., 1883, 10, p. 471.
[2] Centralblatt für klin. Med., 1883, No. 38, p. 609.
[3] Trans. London Path. Soc., 1876, No. 27, p. 148.
[4] Annales de Dermatol. et de Syph., 2d series, 1889, 10, p. 849.
[5] Krankheiten des Kehlkopfes, 1880, p. 362.

disease, as stated above, is by no means rare, Chiari and Riehl,[1] as long ago as 1883, having reported thirty-eight cases of lupus of the larynx. Many more have been reported since.

The following case of primary lupus of the pharynx is, however, the only one that has ever come under my observation. It must be remembered that lupus is one of the diseases that is, judging from literature, much more common in Europe than in this country.

F. F., male, seventeen years of age, an iron-worker, of German parentage, came to the Demilt Dispensary in May, 1890, because his mother, on looking into his throat, had discovered its abnormal appearance. There was no record of hereditary tuberculosis, cancer, or syphilis. The patient himself gave no direct or indirect history of acquired syphilis. About six months previously he began to notice that his throat was dry, his nose was stopped, he breathed through his mouth, and a tickling in his throat made him cough a little. He was slightly deaf on the left side. Three or four months ago he had some lumps removed from the left side of his neck. It was impossible to ascertain whether the appearance of the lumps followed or preceded the throat symptoms, which were exceedingly slight, there being no dysphagia and but little discomfort.

The boy's appetite and general health were good. His appearance was robust; with the exception of the scar at the angle of the jaw on the left side, where enlarged glands could still be indefinitely felt, he presented no external evidence of tuberculous disease. There was no skin-lupus. A physical examination of the chest revealed nothing abnormal. The fauces were seen to be thickened and covered

[1] Vierteljahresschrift für Derm. und Syph., 9, 182, p. 661.

with numerous tiny, shining elevations of the surface. There was some congestion, but no ulceration. The thickening was more marked on the left side. The uvula was enormously hypertrophied, being nearly as large as the distal phalanx of the middle finger. It also was sprinkled with little surface elevations, and near the tip this appearance was exaggerated to such a degree as to be called fungous. There was some reddening around the edges of the infiltrated area on the soft palate, but the tubercles themselves were pale and shining. There was very little secretion. There was some linear cicatricial tissue on the posterior pharyngeal wall. There was no pain on swallowing, or on manipulation. The patient complained only of a clogged feeling in the throat. There was some hypertrophy of the nasal mucous membrane, and some deviation of the septum to the left. It was thought that on the right side a few shining points could be seen on the inferior turbinated bone far back, but at that time this was not satisfactorily established.

After exclusion of syphilis by a thorough but fruitless course of potassium iodide and mercury, the diagnosis of lupus was made. With the cold snare the whole uvula was amputated as close to the velum as possible. Vigorous applications of pure lactic acid were made. The infiltration seemed to subside somewhat, and the patient being relieved of the symptoms that annoyed him, ceased to attend.

A microscopic examination of the excised uvula proved conclusively that we had to deal with a tuberculous growth, notwithstanding that it was impossible to demonstrate the presence of the tubercle-bacillus in the ten or fifteen sections stained for it.

Gottstein,[1] in his excellent work, gives a good description of the microscopic appearances in a case

[1] Krankheiten des Kehlkopfes, 3d ed., p. 277.

of lupus of the uvula, and a reference to his account will give an almost exact picture of my case. Sections were submitted independently to several experienced microscopists, who all agreed as to their tuberculous nature. Several laryngologists saw the case, and the diagnosis of lupus was undisputed.[1]

On the arrival in New York of Koch's fluid—tuberculin—the patient was hunted up and induced to place himself under treatment. Before the inoculations I find in my history-book the following note:

"At the base of the amputated uvula on the anterior inferior edge of the soft palate, there is a rosette of small pin-head, pale, shining tubercles. Here and there a few may be seen along the arch of the soft palate and on the anterior pillar on the left side. The latter is fast to the tonsil, which is hypertrophied. On the posterior pillar on the right side there is a suspicious-looking spot. The epiglottis is thickened, but it is impossible to determine the presence of the little tubercles seen elsewhere; otherwise the larynx is normal in appearance. The patient having had some nose-bleed from the left side, careful examination reveals a little patch of tiny tubercles about the middle of the convex surface of both the middle and inferior turbinated bones on that side. No true ulceration and no destruction of tissue, no breaking down of the epithelium, can be discovered anywhere in the nose or throat."

The patient at that time (December 23d) was

[1] I have also in my possession sections of several specimens of syphilitic disease of the uvula and soft palate, and the contrast between the two forms of inflammation in this locality is striking and interesting.

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shown at the laryngological section of the Academy of Medicine. On December 24, 1890, the patient received his first inoculation with 0.001 of tuberculin, followed at the usual intervals by two or three others. The reaction was characteristic, the fever reaching 101° F., with malaise, etc. The little tubercles that before had been pale-red and shining became white at their centers and looked as if they would soon break down. This, of course, made them much more noticeable, but I am unable to say whether, as Virchow has claimed, new ones were formed, or whether the previously-existing but unnoticeable ones were accentuated and made more evident. There was, of course, a great increase in the congestion; the reddened areola was very marked, and the patient complained of his throat being sore.

He had evidently satisfied himself concerning the effect of the tuberculin and escaped from the hospital. All subsequent attempts to induce him to return were of no avail. In the light of our present experience the folly of his conduct will not appear so glaring as it then did.

The differential diagnosis of syphilis, lupus, and tuberculosis of the mucous membranes—"the three grand processes," as some Frenchman calls them—often presents insurmountable difficulties, even to the most trained and experienced observer, yet typical cases of each are unhesitatingly recognized by the clinician, so unmistakable are their characteristics. The majority of writers claim that there are more cases on the diagnostic border-line between syphilis and lupus than on that between tuberculosis and lupus. I have seen one case of syphilis in which, on the aryteno-epiglottic fold, there was an

appearance almost identical with that described in the case here reported—the same absence of ulceration, the same shining, tiny tubercles. Other manifestations, however, and the result of treatment proved the specific nature of the case.

Clinically, tuberculosis of the throat is a painful, ulcerating, non-cicatrizing affection, almost always combined with visceral tuberculosis, and accompanied by signs of systemic enfeeblement and marked decline of the general health. Lupus, on the contrary, is comparatively painless, does not always tend to the formation of true ulcers, and as it advances leaves cicatricial tissue behind it. As a rule the general health is not affected. It is a matter of dispute whether there is any great preponderance of visceral tuberculosis among its victims, though the weight of evidence, at least in a slight degree, affirms the fact. In considering this question, however, it is always well to remember that about one-third of *all* autopsies show the presence of active or latent tuberculosis, or the evidence of previous tuberculosis. Moreover, although lupus may be followed by tuberculosis in one-half of the instances, it has never been shown that lupus is any more liable to attack the skin or mucous membranes of a consumptive than to attack those of a healthy person.

The writer does not wish to insist on the non-identity of lupus and tuberculosis. He wishes, however, to insist on the Scotch verdict—"not proven." Clinically, we all know the difference in the pictures summoned to the mind of the laryngologist by the words "lupus" and "tuberculosis." The order

of frequency of the occurrence of tuberculosis in the air-passages—and it is of this locality only that the writer pretends to speak—is lungs, larynx, pharynx, nose; while of lupus, it is nose, pharynx, larynx—and lungs never (?).

Histologically, we know how trifling is the difference. The greater vascularity and the fewer areas of coagulation-necrosis, the fewer giant-cells in lupus, are merely matters of degree. The difference in the number of bacilli found in the two processes is, however, too striking not to be significant. In Dr. Rice's recent admirable article[1] on lupus of the throat, there is one point that arrests the attention of any one conversant with the subject, and that is the finding of a considerable number of bacilli in the secretions from the lupus-patch on the face. Through his courtesy I had the privilege of seeing his case, in which the throat lesion differed from that in my case only in the stage of the process and in the location.

The difficulty of demonstrating the tubercle-bacilli in the lupus-tissue is great, it being necessary, usually, to stain many sections before even one bacillus can be made sure of. While I have made no *exhaustive* search for the fact, I have been unable to find in very extensive references *any* record of the bacillus tuberculosis having been found in the secretions of true lupus.[2] L. V. Krynski,[3] how-

[1] New York Med. Record, April 18, 1891.

[2] For an excellent review of this subject, covering in a general way the whole ground, vide Rev. des Sciences Médicales, Avril 15, 1891, No. 74, p. 669.

[3] Deutsche med. Wochenschr., Mai 28, 1891, p. 745.

ever, described a case of facial lupus in the discharges of which he could, as usual, find no tubercle-bacilli; after the first injection of tuberculin, none; after the second, two; after the third injection, countless numbers. After later injections, none were found, except after the fifth, when two were noted. Dr. Rice's paper does not record the relation of the bacterial examination to the time of inoculation.

In the early days of bacteriology the new school had its strongest claim to the belief and confidence of the medical world in the irrefutability of the evidence it presented. Presumptive evidence was ruled out with scorn. We are now asked to accept the tubercle bacillus as the chief, if not the sole factor in the etiology of lupus. No one has ever produced, no one has ever claimed to be able to produce lupus by inoculation of the tubercle-bacillus Tuberculosis is frequently, but not constantly, produced by the inoculation of lupus material; lupus itself has never been reproduced by inoculation. Thus we see that the bacteriological evidence of the identity of the etiology of the two processes is far from satisfactory. We can only suspect that lupus is a variation of tuberculosis. If it should ever be *proved* to be, the term lupus must still be used to indicate a distinct clinical condition.

To ask the observer of diseases of the throat to believe that the two processes are identical in their etiology, is to ask him to lay aside the use of his reason and to disregard the evidence of his senses. He may, however, be disposed to admit the probability of the identity of certain factors in the etiology of each, for disease is not a plant, nor is it an ab-

normal agglomeration of cells in a certain constant order of arrangement; it is "a derangement of any of the vital functions" (Webster). Pathological changes are the results of disease, not the disease itself.

It is perfectly within the range of possibilities, and it is daily becoming more probable, that the same microörganism may cause more diseases than one. It is perfectly certain that we hardly have a disease known to man that is due to any one cause. It is natural for man to seek a simplicity of relation between cause and effect. Such simplicity does not exist between the manifestations, the lesions, and the etiology of disease. Classification of disease is the means, not the end, of its study; and at present no classification is possible that is satisfactory, and it is futile for medical men to try to drag certain clinical manifestations of disease into line with the minute pathological changes with which they are associated. Systems and rules are the crutches with which the medical novice learns to walk, but the sooner he throws them aside when he is on his feet, the less he is encumbered.